Annals of Neurology

Vol 50 July / December 2001

Official Journal of the American Neurological Association and the Child Neurology Society

Editor
Richard T. Johnson, MD
The Johns Hopkins University
Baltimore

Founding Editor, 1977–1984
Fred Plum, MD

Editor, 1985–1992
Arthur K. Asbury, MD

Editor, 1993–1997
Robert A. Fishman, MD

Associate Editors
Ralph W. Kuncl, MD, PhD
Baltimore

David Zee, MD
Baltimore

Associate Editor for Child Neurology
Michael V. Johnston, MD
Baltimore

Managing Editor
Sheila Rose Garrity, MS

Editorial Assistant
Andrea Greise

Publishing Staff
Shawn Morton
Associate Publisher, Medical Sciences

Jennifer Nowak
Senior Production Editor

James G. Pattis, Cunningham Associates
Advertising Representative

Annals of Neurology *(ISSN 0364-5134) is published monthly for the American Neurological Association by Wiley-Liss, Inc.*

Copyright © 2001 Wiley-Liss, Inc. All rights reserved. No part of this publication may be reproduced in any form or by any means, except as permitted under section 107 or 108 of the 1976 United States Copyright Act, without either the prior written permission of the publisher, or authorization through the Copyright Clearance Center, 222 Rosewood Drive, Danvers, MA 01923, (978) 750-8400, fax (978) 750-4470. Requests to the publisher for permission should be addressed to the Permissions Department, John Wiley & Sons, Inc., 605 Third Avenue, New York, NY 10158-0012; (212) 850-6011, fax (212) 850-6008, E-mail: permreq@wiley.com.

Periodicals postage paid at New York, NY, and at additional mailing offices.

Editorial Board
Yves Agid, MD, PhD, Paris
Rose-Mary Boustany, MD, Durham
David R. Cornblath, MD, Baltimore
Stefano Di Donato, MD, Milan
David J. Fink, MD, Pittsburgh
Marc Fisher, MD, Worcester
Kirk A. Frey, MD, PhD, Ann Arbor
Christopher Gomez, MD, Minneapolis
Scott T. Grafton, MD, Hanover, NH
James C. Grotta, MD, Houston
Mark Hallett, MD, Bethesda
Stephen L. Hauser, MD, San Francisco
Bradley T. Hyman, MD, Boston
George Karpati, MD, Montreal
Bruce R. Korf, MD, PhD, Boston
Allan I. Levey, MD, PhD, Atlanta
Henry F. McFarland, MD, Bethesda
Solomon Moshé, MD, New York
Peter C. O'Brien, PhD, Rochester, MN
Alan K. Percy, MD, Birmingham
Ronald C. Petersen, MD, PhD, Rochester, MN
Louis J. Ptáček II, MD, Salt Lake City
Faye S. Silverstein, MD, Ann Arbor
Douglass M. Turnbull, MD, Newcastle Upon Tyne
Ronald J. Tusa, MD, PhD, Atlanta

Annals of Neurology *is indexed in* Index Medicus, Current Contents/Clinical Medicine, Excerpta Medica, Biological Abstracts, Psychological Abstracts; and CINAHL.

Address for subscription information, orders, or changes of address: *John Wiley & Sons, Inc., Customer Service, 605 Third Avenue, New York, NY 10158-0012; (212) 850-6000. If claims are not resolved satisfactorily, please write to Caroline Rothaug, Subscription Fulfillment and Distribution, John Wiley & Sons, Inc., 605 Third Avenue, New York, NY 10158-0012.*

Annual subscription rates: *U.S.: $200.00 individual, $340.00 institution, $89.00 special resident rate; Canada/Mexico: $200.00 individual, $340.00 institution. All other countries: $269.00 individual, $469.00 institution. Single copies may be ordered from the publisher for $39.00 (add $5.00 for postage outside U.S.). Subscription Price: (Volumes 51 and 52, 2002) Print only: $370.00 in U.S., $370.00 in Canada and Mexico, $499.00 outside North America. Electronic only: $370.00 worldwide. A combination price of $389.00 in U.S., $389.00 in Canada and Mexico, $518.00 outside North America, includes the subscription in both electronic and print formats. Personal rate: $215.00 in North America, and $284.00 rest of world. All subscriptions containing a print element, shipped outside U.S., will be sent by air. Payment must be made in U.S. dollars drawn on a U.S. bank. Claims for undelivered copies will be accepted only after the following issue has been delivered. Prices subject to change without notice. Copies will be replaced without charge if the publisher receives a request within 90 days of the mailing date, both in the U.S. and worldwide.*

POSTMASTER: *Send address changes to* Annals of Neurology, *Subscription Fulfillment and Distribution, John Wiley & Sons, Inc., 605 Third Avenue, New York, NY 10158-0012.*

WILEY-LISS

NO 1 JULY 2001

Editorial

1 **Annals 25th Anniversary**
Fred Plum

Notice

4 **Ad Hoc Reviewers**

Neurological Progress

6 **Neuroimmunophilins: Novel Neuroprotective and Neuroregenerative Targets**
Xin Guo, PhD, James F. Dillman III, PhD, Valina L. Dawson, PhD, and Ted M. Dawson, MD, PhD

Original Articles

17 **Small Fibers Involvement in Friedreich's Ataxia**
Maria Nolano, MD, Vincenzo Provitera, MD, Claudio Crisci, MD, Anna Maria Saltalamacchia, TN, Gwen Wendelschafer-Crabb, MS, William Robert Kennedy, MD, Alessandro Filla, MD, Lucio Santoro, MD, and Giuseppe Caruso, MD

26 **Sodium Channel Inactivation in an Animal Model of Acute Quadriplegic Myopathy**
Mark M. Rich, MD, PhD, and Martin J. Pinter, PhD

34 **Subclinical Dopaminergic Dysfunction in Asymptomatic Parkinson's Disease Patients' Relatives with a Decreased Sense of Smell**
Henk W. Berendse, MD, PhD, Jan Booij, MD, PhD, Chantal M. J. E. Francot, MSc, Paul L. M. Bergmans, MSc, Ron Hijman, PhD, Johannes C. Stoof, PhD, and Erik Ch. Wolters, MD, PhD

42 **Diagnosis of X-Linked Myotubular Myopathy by Detection of Myotubularin**
Jocelyn Laporte, PhD, Wolfram Kress, MD, PhD, and Jean-Louis Mandel, MD, PhD

47 **PMP22 Transgenic Dorsal Root Ganglia Cultures Show Myelin Abnormalities Similar to Those of Human CMT1A**
Lucilla Nobbio, PhD, Gianluigi Mancardi, MD, Marina Grandis, MD, Giovanni Levi, PhD, Ueli Suter, PhD, Klaus-Armin Nave, PhD, Anthony J. Windebank, MD, Michele Abbruzzese, MD, and Angelo Schenone, MD

56 **Prospective Study of Caffeine Consumption and Risk of Parkinson's Disease in Men and Women**
Alberto Ascherio, MD, DrPH, Shumin M. Zhang, MD, ScD, Miguel A. Hernán, MD, DrPH, Ichiro Kawachi, MD, PhD, Graham A. Colditz, MD, DrPH, Frank E. Speizer, MD, and Walter C. Willett, MD, DrPH

64 **Mature, Long-lived $CD4^+$ and $CD8^+$ T Cells Are Generated by the Thymoma in Myasthenia Gravis**
Camilla Buckley, MRCP, Daniel Douek, PhD, John Newsom-Davis, FRS, Angela Vincent, FRCPath, and Nicholas Willcox, PhD

73 **Potassium Channel Antibodies in Two Patients with Reversible Limbic Encephalitis**
Camilla Buckley, MD, Joel Oger, MD, Linda Clover, BSc, Erdem Tüzün, MD, Katherine Carpenter, Dip Psych, Matthew Jackson, MD, and Angela Vincent, FRCPath

79 **Brain Magnetic Resonance Imaging in 23 Patients with Mucopolysaccharidoses and the Effect of Bone Marrow Transplantation**
Toshiyuki Seto, MD, Kinuko Kono, MD, Kyoko Morimoto, MD, Yuichi Inoue, MD, Haruo Shintaku, MD, Hideji Hattori, MD, Osamu Matsuoka, MD, Tsunekazu Yamano, MD, and Akemi Tanaka, MD

93 **Microarray Analysis of Hippocampal Gene Expression in Global Cerebral Ischemia**
Kunlin Jin, MD, PhD, Xiao Ou Mao, MD, Mark W. Eshoo, PhD, Tetsuya Nagayama, MD, Manabu Minami, MD, Roger P. Simon, MD, and David A. Greenberg, MD, PhD

Brief Communications

104 **Progressive Mitochondrial Disease Resulting from a Novel Missense Mutation in the Mitochondrial DNA ND3 Gene**
Robert W. Taylor, PhD, Rajinder Singh-Kler, PhD, Christine M. Hayes, BSc, Philip E.M. Smith, FRCP, and Douglass M. Turnbull, FRCP

108 **Late-Onset Metachromatic Leukodystrophy Clinically Presenting as Isolated Peripheral Neuropathy: Compound Heterozygosity for the IVS2+1G→A Mutation and a Newly Identified Missense Mutation (Thr408Ile) in a Spanish Family**
Manuel Comabella, MD, John S. Waye, PhD, Nuria Raguer, MD, Barry Eng, BSc, Carmen Domínguez, PhD, Carmen Navarro, MD, Cecilia Borrás, PhD, William Krivit, MD, PhD, and Xavier Montalbán, MD

112 **Dichloroacetate Exerts Therapeutic Effects in Transgenic Mouse Models of Huntington's Disease**
Ole A. Andreassen, MD, Robert J. Ferrante, PhD, Hsueh-Meei Huang, PhD, Alpaslan Dedeoglu, PhD, Larry Park, PhD, Kimberly L. Ferrante, BA, Jennifer Kwon, BS, David R. Borchelt, PhD, Christopher A. Ross, MD, PhD, Gary E. Gibson, PhD, and M. Flint Beal, MD

117 **Familial Frontotemporal Dementia and Parkinsonism with a Novel Mutation at an Intron 10+11-Splice Site in the *tau* Gene**
Katsuichi Miyamoto, MD, PhD, Anna Kowalska, PhD, Masato Hasegawa, PhD, Takeshi Tabira, MD, PhD, Keikichi Takahashi, PhD, Wataru Araki, MD, PhD, Ichiro Akiguchi, MD, PhD, and Akito Ikemoto, MD, PhD

Special Report

121 **Recommended Diagnostic Criteria for Multiple Sclerosis: Guidelines from the International Panel on the Diagnosis of Multiple Sclerosis**
W. Ian McDonald, FRCP, Alistair Compston, FRCP, Gilles Edan, MD, Donald Goodkin, Hans-Peter Hartung, MD, Fred D. Lublin, MD, Henry F. McFarland, MD, Donald W. Paty, MD, Chris H. Polman, MD, Stephen C. Reingold, PhD, Magnhild Sandberg-Wollheim, MD, William Sibley, MD, Alan Thompson, MD, Stanley van den Noort, MD, Brian Y. Weinshenker, MD, and Jerry S. Wolinsky, MD

Letters

128 **Increased Hair Polyamine Levels in Patients with Alzheimer's Disease**
Man Ho Choi, MSc, Kyoung-Rae Kim, PhD, In Seong Kim, MD, Dong Seok Lho, PhD, and Bong Chul Chung, PhD

128 **Hiccups Start, Seizures Cease**
Akio Ikeda, MD, PhD, Keiko Matsuoka, MD, Yoshihumi Nakaya, MD, and Hiroshi Shibasaki, MD, PhD

Erratum

129 **Reply**
Christophe Tzourio, MD, PhD

Regular Departments

130 **Books**

NO 2 AUGUST 2001

Original Articles

133 **Aerobic Conditioning in Patients with Mitochondrial Myopathies: Physiological, Biochemical, and Genetic Effects**
Tanja Taivassalo, PhD, Eric A. Shoubridge, PhD, Jacqueline Chen, M Eng, Nancy G. Kennaway, DPhil, Salvatore DiMauro, MD, Douglas L. Arnold, MD, and Ronald G. Haller, MD

142 **Metabolic Enzyme Expression in Dopaminergic Neurons in Parkinson's Disease: An In Situ Hybridization Study**
Ann E. Kingsbury, Mark Cooper, PhD, Anthony H. V. Schapira, DSc, MD, FRCP, and Oliver J. F. Foster, PhD, FRCP

150 **Large-Scale, Multicenter Study of Cerebrospinal Fluid Tau Protein Phosphorylated at Serine 199 for the Antemortem Diagnosis of Alzheimer's Disease**
Nobuo Itoh, MD, Hiroyuki Arai, MD, PhD, Katsuya Urakami, MD, Koichi Ishiguro, PhD, Hideto Ohno, PhD, Harald Hampel, MD, Katharina Buerger, MD, Jens Wiltfang, MD, Markus Otto, MD, Hans Kretzschmar, MD, Hans-Juergen Moeller, MD, Masaki Imagawa, MD, Hideki Kohno, PhD, Kenji Nakashima, MD, Shigeki Kuzuhara, MD, Hidetada Sasaki, MD, PhD, and Kazutomo Imahori, PhD

157 **Increased Frequency of the Tumor Necrosis Factor-α-308 A Allele in Adults with Human Immunodeficiency Virus Dementia**
Michael W. Quasney, MD, PhD, Qing Zhang, BS, Susie Sargent, MD, Margaret Mynatt, BS, Jonathan Glass, MD, and Justin McArthur, MD

163 **Presumed Pre- or Perinatal Arterial Ischemic Stroke: Risk Factors and Outcomes**
Meredith R. Golomb, MD, Daune L. MacGregor, MD, Trish Domi, Derek C. Armstrong, MD, Brian W. McCrindle, MD, Supriya Mayank, MD, and Gabrielle A. deVeber, MD

169 **Multiple Sclerosis: Altered Glutamate Homeostasis in Lesions Correlates with Oligodendrocyte and Axonal Damage**
Peter Werner, PhD, David Pitt, MD, and Cedric S. Raine, PhD

181 **Blinded Positron Emission Tomography Study of Dopamine Cell Implantation for Parkinson's Disease**
Toshitaka Nakamura, MD, Vijay Dhawan, PhD, Thomas Chaly, PhD, Masafumi Fukuda, MD, Yilong Ma, PhD, Robert Breeze, MD, Paul Greene, MD, Stanley Fahn, MD, Curt Freed, MD, and David Eidelberg, MD

188 **Brain-Derived Neurotrophic Factor Enhances Fast Excitatory Synaptic Transmission in Human Epileptic Dentate Gyrus**
Wei Jian Zhu, MD, PhD, and Steven N. Roper, MD

195 **Randomized Controlled Trial of Intravenous Immunoglobulin Versus Oral Prednisolone in Chronic Inflammatory Demyelinating Polyradiculoneuropathy**
Richard Hughes, MD, FMedSci, Siân Bensa, BSc, Hugh Willison, PhD, Peter Van den Bergh, MD, PhD, Giancarlo Comi, PhD, Isabel Illa, MD, PhD, Eduardo Nobile-Orazio, PhD, Pieter van Doorn, PhD, Marinos Dalakas, MD, Martin Bojar, CSc, Anthony Swan, PhD, and the Inflammatory Neuropathy Cause and Treatment (INCAT) Group

202 **β-Enolase Deficiency, a New Metabolic Myopathy of Distal Glycolysis**
Giacomo P. Comi, MD, Francesco Fortunato, BS, Sabrina Lucchiari, PhD, Andreina Bordoni, BS, Alessandro Prelle, MD, Stefano Jann, MD, Angélica Keller, PhD, Patrizia Ciscato, BS, Sara Galbiati, MD, Luca Chiveri, MD, Yvan Torrente, MD, Guglielmo Scarlato, MD, and Nereo Bresolin, MD

208 **Imaging Appearance of the Symptomatic Perforating Artery in Patients with Lacunar Infarction: Occlusion or Other Vascular Pathology?**
Joanna M. Wardlaw, FRCR, FRCP, MD, Martin S. Dennis, FRCP, MD, Charles P. Warlow, FRCP, MD, and Peter A. Sandercock, FRCP, MD

216 **Chromogranins as Markers of Altered Hippocampal Circuitry in Temporal Lobe Epilepsy**
Susanne Pirker, MD, Thomas Czech, MD, Christoph Baumgartner, MD, Hans Maier, MD, Klaus Novak, MD, Sabine Fürtinger, MSc, Reiner Fischer-Colbrie, PhD, and Günther Sperk, PhD

227 **Increased Incidence of Sporadic Creutzfeldt-Jakob Disease on the Island of Crete Associated with a High Rate of *PRNP* 129-Methionine Homozygosity in the Local Population**
Andreas Plaitakis, MD, Anna K. Viskadouraki, MD, Minas Tzagournissakis, MD, Ioannis Zaganas, MD, Susan Verghese-Nikolakaki, PhD, Vasilis Karagiorgis, Ioannis Panagiotides, MD, Constantine Kilindireas, MD, Eustratios Patsouris, MD, Christine Haberler, MD, Herbert Budka, MD, and Theodoros Sklaviadis, PhD

234 **Clinical and Genetic Analysis of Four Mexican Families With Spinocerebellar Ataxia Type 10**
Astrid Rasmussen, MD, Tohru Matsuura, MD, Luis Ruano, MD, Petra Yescas, MSc, Adriana Ochoa, BA, Tetsuo Ashizawa, MD, and Elisa Alonso, MD

240 **Reorganization of Human Motor Cortex After Hand Replantation**
Simone Röricht, MD, Jochen Machetanz, MD, Kerstin Irlbacher, MD, Ludwig Niehaus, MD, Edgar Biemer, MD, and Bernd-Ulrich Meyer, MD

Brief Communications

250 **Nonprogressive Autosomal Recessive Ataxia Maps to Chromosome 9q34-9qter in a Large Consanguineous Lebanese Family**
Valérie Delague, PhD, Corinne Bareil, PhD, Patrice Bouvagnet, MD, PhD, Nabiha Salem, MA, Eliane Chouery, MA, Jacques Loiselet, MD, PhD, André Mégarbané, MD, PhD, and Mireille Claustres, MD, PhD

254 **Levodopa Induces Dyskinesias in Normal Squirrel Monkeys**
Daniel M. Togasaki, MD, PhD, Louis Tan, MRCP, Peter Protell, BA, Donato A. Di Monte, MD, Maryka Quik, PhD, and J. William Langston, MD

258 **Sporadic Creutzfeldt-Jakob Disease in a Young Dutch Valine Homozygote: Atypical Molecular Phenotype**
Mark W. Head, PhD, Gerrit Tissingh, MD, Bernard M. J. Uitdehaag, MD, PhD, Frederik Barkhof, MD, PhD, Tristan J. R. Bunn, MSc, James W. Ironside, FRCP, Wouter Kamphorst, MD, PhD, and Philip Scheltens, MD, PhD

261 **Frameshift Mutation in the Collagen VI Gene Causes Ullrich's Disease**
Itsuro Higuchi, MD, Tadafumi Shiraishi, MD, Teruto Hashiguchi, MD, Masahito Suehara, MD, Takahito Niiyama, MD, Masanori Nakagawa, MD, Kimiyoshi Arimura, MD, Ikuro Maruyama, MD, and Mitsuhiro Osame, MD

266 **Proton Magnetic Resonance Spectroscopy of Brain Metabolites in Galactosemia**
Zhiyue J. Wang, PhD, Gerard T. Berry, MD, Steffi F. Dreha, MD, Huaqing Zhao, MA, Stanton Segal, MD, and Robert A. Zimmerman, MD

269 **Adult Neuronal Ceroid Lipofuscinosis with Palmitoyl-Protein Thioesterase Deficiency: First Adult-Onset Patients of a Childhood Disease**
Otto P. van Diggelen, PhD, Stephane Thobois, MD, Caroline Tilikete, MD, Marie-Thérèse Zabot, PhD, Joke L. M. Keulemans, BS, Patrick A. van Bunderen, BS, Peter E. M. Taschner, PhD, Monique Losekoot, PhD, and Yakov V. Voznyi, PhD

Letters

273 **Survival in a Transgenic Model of FALS Is Independent of iNOS Expression**
Marjatta Son, PhD, Hassan M. Fathallah-Shaykh, MD, and Jeffrey L. Elliott, MD

273 **Herpes Simplex Virus Type 1-Induced Acute Retinal Necrosis**
Kei Numazaki, MD, PhD, and Norihiko Itoh, DVM

274 **Reply**
Georges M. G. M. Verjans, PhD, and Albert D. M. E. Osterhaus, DVM, PhD

275 **Interleukin-12p40 Genotype Plays a Role in the Susceptibility to Multiple Sclerosis**
Tineke van Veen, MSc, J. Bart A. Crusius, MSc, Hans M. Schrijver, MD, Gerd Bouma, MD, PhD, Joep Killestein, MD, Lisa van Winsen, MD, A. Salvador Peña, MD, PhD, Chris H. Polman, MD, PhD, and Bernard M.J. Uitdehaag, MD, PhD

275 **Polymorphism of the Interleukin-1 Gene Complex in Localization-Related Epilepsy**
Jukka Peltola, MD, Tapani Keränen, MD, PhD, Sirpa Rainesalo, MD, and Mikko Hurme, MD, PhD

Regular Departments

277 **Books**

Editorials

281 **Chronic Inflammatory Demyelinating Polyradiculoneuropathy**
Richard A. C. Hughes, MD, FRCP, FMedSci

283 **Parkin and Parkinson's: More than Homonymy?**
Olga Corti, PhD, and Alexis Brice, MD

Original Articles

286 **P0 Protein Is a Target Antigen in Chronic Inflammatory Demyelinating Polyradiculoneuropathy**
Wei Xing Yan, MD, Juan J. Archelos, MD, Hans-Peter Hartung, MD, and John D. Pollard, BSc, MD, PhD

293 **Lewy Bodies and Parkinsonism in Families with *Parkin* Mutations**
Matt Farrer, PhD, Piu Chan, MD, Rong Chen, PhD, Louis Tan, MD, Sarah Lincoln, BS, Dena Hernandez, BS, Lysia Forno, MD, Katrina Gwinn-Hardy, MD, Leonard Petrucelli, PhD, Jennifer Hussey, BS, Andrew Singleton, PhD, Caroline Tanner, MD, PhD, John Hardy, PhD, and J. William Langston, MD

301 **ANNA-3 Anti-Neuronal Nuclear Antibody: Marker of Lung Cancer-Related Autoimmunity**
Koon Ho Chan, MD, Steven Vernino, MD, PhD, and Vanda A. Lennon, MD, PhD

312 **Nemaline Myopathy: A Clinical Study of 143 Cases**
Monique M. Ryan, MB, BS, Christina Schnell, MS, Corinne D. Strickland, MS, Lloyd K. Shield, MB, BS, Graeme Morgan, MB, BS, Susan T. Iannaccone, MD, Nigel G. Laing, PhD, Alan H. Beggs, PhD, and Kathryn N. North, MD, BS

321 **Nigrostriatal Collaterals to Thalamus Degenerate in Parkinsonian Animal Models**
Amanda Freeman, BS, Brian Ciliax, PhD, Roy Bakay, MD, Joseph Daley, BS, R. Daniel Miller, MS, Glenda Keating, PhD, Allan Levey, MD, PhD, and David Rye, MD, PhD

330 **Central and Peripheral Nervous System Infection, Immunity, and Inflammation in the NHP Model of Lyme Borreliosis**
Andrew R. Pachner, MD, Diego Cadavid, MD, Gale Shu, BS, Donna Dail, MS, Sarah Pachner, Emir Hodzic, DVM, and Stephen W. Barthold, DVM, PhD

339 **Molecular and Clinical Diversity in Paraneoplastic Immunity to Ma Proteins**
Myrna R. Rosenfeld, MD, PhD, Joseph G. Eichen, MS, Deborly F. Wade, BS, Jerome B. Posner, MD, and Josep Dalmau, MD, PhD

349 **Complex Immunomodulatory Effects of Interferon-β in Multiple Sclerosis Include the Upregulation of T Helper 1-Associated Marker Genes**
Klaus-Peter Wandinger, MD, Claus-Steffen Stürzebecher, MD, Bibiana Bielekova, MD, Greg Detore, Andreas Rosenwald, MD, Louis M. Staudt, MD, PhD, Henry F. McFarland, MD, and Roland Martin, MD

358 **Alzheimer's Disease versus Dementia with Lewy Bodies: Cerebral Metabolic Distinction with Autopsy Confirmation**
Satoshi Minoshima, MD, PhD, Norman L. Foster, MD, Anders A. F. Sima, MD, PhD, Kirk A. Frey, MD, PhD, Roger L. Albin, MD, and David E. Kuhl, MD

366 **Topiramate Blocks Perinatal Hypoxia-Induced Seizures in Rat Pups**
Sookyong Koh, MD, PhD, and Frances E. Jensen, MD

373 **A Disorder Similar to Huntington's Disease Is Associated with a Novel CAG Repeat Expansion**
Russell L. Margolis, MD, Elizabeth O'Hearn, MD, Adam Rosenblatt, MD, Virginia Willour, PhD, Susan E. Holmes, PhD, Mary L. Franz, MSW, Colleen Callahan, BA, Hyon S. Hwang, BA, Juan C. Troncoso, MD, and Christopher A. Ross, MD, PhD

381 **Low Cerebrospinal Fluid Hypocretin (Orexin) and Altered Energy Homeostasis in Human Narcolepsy**
Seiji Nishino, MD, PhD, Beth Ripley, BS, Sebastiaan Overeem, MSc, Sona Nevsimalova, MD, PhD, Gert Jan Lammers, MD, PhD, Jitka Vankova, MD, Michele Okun, MA, William Rogers, BS, Stephen Brooks, MD, and Emmanuel Mignot, MD, PhD

389 **Transected Neurites, Apoptotic Neurons, and Reduced Inflammation in Cortical Multiple Sclerosis Lesions**
John W. Peterson, BS, Lars Bö, MD, PhD, Sverre Mörk, MD, PhD, Ansi Chang, MD, and Bruce D. Trapp, PhD

Brief Communications

401 **Pure Representational Neglect After Right Thalamic Lesion**
Stéphanie Ortigue, MS, Isabelle Viaud-Delmon, PhD, Jean-Marie Annoni, MD, Theodor Landis, MD, Christoph Michel, PhD, Olaf Blanke, MD, Patrik Vuilleumier, MD, and Eugène Mayer, PhD

404 **Elevated Corticotropin Releasing Hormone/ Corticotropin Releasing Hormone-R1 Expression in Postmortem Brain Obtained from Children with Generalized Epilepsy**
Wei Wang, MD, Kimberly E. Dow, MD, and Douglas D. Fraser, MD, PhD,

409 **Early-Onset Multisystem Mitochondrial Disorder Caused by a Nonsense Mutation in the Mitochondrial DNA *Cytochrome C Oxidase II* Gene**
Yolanda Campos, MSc, Alberto García-Redondo, MSc, Miguel A. Fernández-Moreno, PhD, Mercedes Martínez-Pardo, MD, Guillermo Goda, Pharm B, Juan C. Rubio, MSc, Miguel A. Martín, Pharm B, Pilar del Hoyo, Pharm B, Ana Cabello, MD, Belen Bornstein, MD, Rafael Garesse, PhD, and Joaquín Arenas, PhD

413 **Ocular Flutter Associated with a Localized Lesion in the Paramedian Pontine Reticular Formation**
Fred Schon, FRCP, Timothy L. Hodgson, PhD, Dominic Mort, MRCP, and Christopher Kennard, FRCP

Expedited Publication

417 **Sodium Channel Inactivation Defects Are Associated with Acetazolamide-Exacerbated Hypokalemic Periodic Paralysis**
Saïd Bendahhou, PhD, Theodore R. Cummins, PhD, Robert C. Griggs, MD, Ying-Hui Fu, PhD, and Louis J. Ptácek, MD

Letters

421 **Alteration of the Serotoninergic System in Fatal Familial Insomnia**
Pietro Cortelli, Ronald Polinsky, Pasquale Montagna, and Elio Lugaresi

421 **Reply**
Julia Wanschitz, MD, Marin Guentchev, MD, and Herbert Budka, MD

422 **Effects of Seasons on Magnetic Resonance Imaging-Measured Disease Activity in Patients with Multiple Sclerosis**
Helen Tremlett, BPharm Hons, PhD

422 **Reply**
Marco Rovaris, MD, Giancarlo Comi, MD, Maria Pia Sormani, PhD, Jerry S. Wolinsky, MD, David Ladkani, PhD, and Massimo Filippi, MD

423 **Borna Disease Virus RNA Is Absent in Chronic Multiple Sclerosis**
Claus G. Haase, MD, Sergei Viazov, MD, Melanie Fiedler, MD, Nikolaus Koenig, MD, Pedro M. Faustmann, MD, and Michael Roggendorf, MD

424 **Monozygotic Twins with X-Linked Adrenoleukodystrophy and Different Phenotypes**
Maja Di Rocco, MD, Laura Doria-Lamba, MD, and Ubaldo Caruso, PhD

Regular Departments

425 **Books**

NO 3 SEPTEMBER 2001 SUPPLEMENT 1

126th Annual Meeting of the American Neurological Association

S1 **Program**
S8 **Abstracts**

Neurology Outcomes Research: Current Science and Future Directions

S67 **Program**
S69 **Abstracts**

30th Annual Meeting of the Child Neurology Society

S80 **Program**
S92 **Abstracts**

NO 4 OCTOBER 2001

Editorials

429 **Endogenous Retroviruses and Multiple Sclerosis**
Dennis L. Kolson, MD, PhD, and Francisco González-Scarano, MD

431 **Matrix Metalloproteinases in Multiple Sclerosis: Is It Time for a Treatment Trial?**
Gary A. Rosenberg, MD

Original Articles

434 **Monocyte Activation and Differentiation Augment Human Endogenous Retrovirus Expression: Implications for Inflammatory Brain Diseases**
James B. Johnston, PhD, Claudia Silva, MSc, Janet Holden, MD, Kenneth G. Warren, MD, Arthur W. Clark, MD, and Christopher Power, MD

443 **Matrix Metalloproteinases and Their Tissue Inhibitors as Markers of Disease Subtype and Response to Interferon-β Therapy in Relapsing and Secondary-Progressive Multiple Sclerosis Patients**
Yanina Galboiz, MSc, Sarah Shapiro, PhD, Nitza Lahat, PhD, Hannah Rawashdeh, MD, and Ariel Miller, MD, PhD

452 **Hereditary Motor and Sensory Neuropathy-Russe: New Autosomal Recessive Neuropathy in Balkan Gypsies**
P. K. Thomas, DSc, MD, Luba Kalaydjieva, PhD, MD, Brian Youl, MD, Tamara Rogers, PhD, Dora Angelicheva, PhD, Rosalind H. M. King, PhD, Velina Guergueltcheva, MD, Jaume Colomer, MD, Constantin Lupu, MD, Axinia Corches, MD, Gabriela Popa, MD, Luciano Merlini, MD, Alex Shmarov, PhD, MD, John R. Muddle, Michelle Nourallah, and Ivailo Tournev, PhD, MD

458 **Are Certain Diuretics Also Anticonvulsants?**
Dale C. Hesdorffer, PhD, James P. Stables, MSA, W. Allen Hauser, MD, John F. Annegers, PhD, and Gregory Cascino, MD

463 **Individual Blood–Brain Barrier Phenylalanine Transport Determines Clinical Outcome in Phenylketonuria**
Josef Weglage, MD, PhD, Dirk Wiedermann, PhD, Jonas Denecke, MD, Reinhold Feldmann, PhD, Hans-Georg Koch, MD, Kurt Ullrich, MD, Erik Harms, MD, and Harald E. Möller, PhD

468 **Human Apolipoprotein E4 Accelerates β-Amyloid Deposition in APPsw Transgenic Mouse Brain**
Donald B. Carter, PhD, Edwige Dunn, MS, Denise D. McKinley, BS, Nancy C. Stratman, MS, Timothy P. Boyle, BS, Susan L. Kuiper, MS, Jo A. Oostveen, MS, Royal J. Weaver, PhD, Jennifer A. Boller, BS, and Mark E. Gurney, PhD

476 **Autosomal Dominant Glut-1 Deficiency Syndrome and Familial Epilepsy**
Knut Brockmann, MD, Dong Wang, MD, Christoph G. Korenke, MD, Arpad von Moers, MD, Yuan-Yuan Ho, PhD, Juan M. Pascual, MD, PhD, Kunyan Kuang, MD, Hong Yang, MD, Li Ma, Pamela Kranz-Eble, BS Jorge Fischbarg, MD, PhD, Folker Hanefeld, MD, and Darryl C. De Vivo, MD

486 **Elevated Serum and Cerebrospinal Fluid Levels of Soluble Human Herpesvirus Type 6 Cellular Receptor, Membrane Cofactor Protein, in Patients with Multiple Sclerosis**
Samantha S. Soldan, MS, Anna Fogdell-Hahn, PhD, Meghan B. Brennan, BA, Barbara B. Mittleman, MD, Clara Ballerini, PhD, Luca Massacesi, MD, Tsukasa Seya, MD, PhD, Henry F. McFarland, MD, and Steven Jacobson, PhD

494 **Relationship of the Extended Tau Haplotype to Tau Biochemistry and Neuropathology in Progressive Supranuclear Palsy**
Wan-Kyng Liu, PhD, Tien V. Le, BS, Jennifer Adamson, BS, Matthew Baker, BSc, Natalie Cookson, BS, John Hardy, PhD, Michael Hutton, PhD, Shu-Hui Yen, PhD, and Dennis W. Dickson, MD

503 **Pick's Disease Associated with the Novel *Tau* Gene Mutation K369I**
Manuela Neumann, MD, Walter Schulz-Schaeffer, MD, R. Anthony Crowther, PhD, Michael J. Smith, Maria Grazia Spillantini, PhD, Michel Goedert, MD, PhD, and Hans A. Kretzschmar, MD

514 **Metabolic Changes Following Subthalamotomy for Advanced Parkinson's Disease**
Philip C. Su, MD, Yilong Ma, PhD, Masafumi Fukuda, MD, Marc J. Mentis, MD, Ham-Min Tseng, MD, Ruoh-Fang Yen, MD, Hon-Man Liu, MD, J. R. Moeller, PhD, and David Eidelberg, MD

521 **Human Brain Mapping in Dystonia Reveals Both Endophenotypic Traits and Adaptive Reorganization**
Sabine Meunier, MD, PhD, Line Garnero, PhD, Antoine Ducorps, BSc, Leonor Mazières, MD, PhD, Stéphane Lehéricy, MD, PhD, Sophie Tézenas du Montcel, MD, Bernard Renault, PhD, and Marie Vidailhet, MD, PhD

Brief Communications

528 **Neuronal Activity in the Globus Pallidus in Chorea Caused by Striatal Lacunar Infarction**
Takao Hashimoto, MD, PhD, Hiroshi Morita, MD, PhD, Tsuyoshi Tada, MD, PhD, Tetsuhiro Maruyama, MD, PhD, Yuzo Yamada, MD, PhD, and Shu-ichi Ikeda, MD, PhD

532 **Myelinopathia Centralis Diffusa (Vanishing White Matter Disease): Evidence of Apoptotic Oligodendrocyte Degeneration in Early Lesion Development**
Wolfgang Brück, MD, Jochen Herms, MD, Knut Brockmann, MD, Walter Schulz-Schaeffer, MD, and Folker Hanefeld, MD

537 **A Novel Deafness/Dystonia Peptide Gene Mutation that Causes Dystonia in Female Carriers of Mohr-Tranebjaerg Syndrome**
Russell H. Swerdlow, MD, and G. Frederick Wooten, MD

540 **Multisystem Disorder Associated with a Missense Mutation in the Mitochondrial Cytochrome *b* Gene**
Flemming Wibrand, PhD, Kirstine Ravn, MSc, Marianne Schwartz, PhD, Thomas Rosenberg, MD, Nina Horn, PhD, and John Vissing, MD, PhD

544 **Dextran Reduces Embolic Signals After Carotid Endarterectomy**
Christopher R. Levi, MBBS, FRACP, Jacinda L. Stork, BA, BAppSci (Hons), Brian R. Chambers, MD, FRACP, Anne L. Abbott, MBBS, FRACP, Heather M. Cameron, RN, Anna Peeters, BSc (Hons), PhD, John P. Royle, FRACS, FACS, Andrew K. Roberts, FRACS, FACS, Gary Fell, MBBS, FRACS, Michael C. Hoare, MBBS, FRACS, Anthony T. W. Chan, Mmed (Surg), FRACS, and Geoffrey A. Donnan, MD, FRACP

547 **Mutation of the Doublecortin Gene in Male Patients with Double Cortex Syndrome: Somatic Mosaicism Detected by Hair Root Analysis**
Mitsuhiro Kato, MD, Masayo Kanai, MD, Osamu Soma, MD, Yuichi Takusa, MD, Toshiyuki Kimura, MD, Chikahiko Numakura, MD, Takasumi Matsuki, MD, Shigeki Nakamura, PhD, and Kiyoshi Hayasaka, MD

Letters

552 **Conjugal Multiple Sclerosis**
Christopher Hawkes, MD, FRCP

552 **Reply**
George C. Ebers, MD, A. D. Sadovnick, PhD, FRCP(c), and Pierre Duquette, FRCP(c), for the Canadian Collaborative Study

553 **Compound Heterozygosity and Variable Penetrance in *SOD1* Amyotrophic Lateral Sclerosis Pedigrees**
Matthew J. Parton, MRCP, Peter M. Andersen, MD, DMSc, Wendy J. Broom, BSc, and Christopher E. Shaw, MD

554 **Reply**
Collette K. Hand, PhD, Veronique Mayeux-Portas, PhD, Jawad Khoris, MD, Valerie Briolotti, Pierre Clavelou, MD, PhD, William Camu, MD, PhD, and Guy A. Rouleau, MD, PhD

554 **Electroencephalographic and Clinical Correlation of Hyponatremia Induced During Transurethral Resection of the Prostate**
Venkata R. Reddy, MD, and Sreenivasa S. Moorthy, MD

Obituary

556 **Clarence Joseph Gibbs, Jr, 1924–2001**
Richard T. Johnson, MD

Regular Departments

557 **Books**

NO 5 NOVEMBER 2001

Editorial

559 **Behavioral Neurology in the Emergency Room: Language Testing, Brain Imaging, and Acute Stroke Therapy**
Howard S. Kirshner, MD

Original Articles

561 **Hypoperfusion of Wernicke's Area Predicts Severity of Semantic Deficit in Acute Stroke**
Argye E. Hillis, MD, MS, Robert J. Wityk, MD, Elizabeth Tuffiash, Norman J. Beauchamp, MD, MSH, Michael A. Jacobs, PhD, Peter B. Barker, DPhil, and Ola A. Selnes, PhD

567 **Attention and Cognition in Bradykinetic–Rigid Syndromes: An Event-Related Potential Study**
Zvezdan Pirtošek, MD, Marjan Jahanshahi, PhD, Geoff Barrett, PhD, and Andrew J. Lees, MD, FRCP

574 **Molecular Heterogeneity of Myophosphorylase Deficiency (McArdle's Disease): A Genotype–Phenotype Correlation Study**
Miguel A. Martín, Pharm B, Juan C. Rubio, MSc, Jenny Buchbinder, PhD, Roberto Fernández-Hojas, PhD, Pilar del Hoyo, Pharm B, Susana Teijeira, PhD, Josep Gámez, MD, Carmen Navarro, MD, José M. Fernández, MD, Ana Cabello, MD, Yolanda Campos, MSc, Carlos Cervera, MD, José M. Culebras, PhD, Antoni L. Andreu, PhD, Robert Fletterick, PhD, and Joaquín Arenas, PhD

582 **Magnetoencephalographic Fields from Patients with Spontaneous and Induced Migraine Aura**
Susan M. Bowyer, PhD, Sheena K. Aurora, MD, John E. Moran, PhD, Norman Tepley, PhD, and K. M. A. Welch, MD

588 **Poststreptococcal Acute Disseminated Encephalomyelitis with Basal Ganglia Involvement and Auto-Reactive Antibasal Ganglia Antibodies**
Russell C. Dale, MRCP, Andrew J. Church, BSc, Francisco Cardoso, PhD, Elizabeth Goddard, Tim C. Cox, W. K. "Kling" Chong, Amanda Williams, MRCP, Nigel J. Klein, PhD, Brian G. Neville, FRCP, Edward J. Thompson, DSc, and Gavin Giovannoni, PhD

596 **Sexual Dimorphism and Developmental Regulation of Substantia Nigra Function**
Jana Velíšková, MD, PhD, and Solomon L. Moshé, MD

602 **A Nitric Oxide Donor Induces Neurogenesis and Reduces Functional Deficits After Stroke in Rats**
Ruilan Zhang, MD, Li Zhang, MD, Zhenggang Zhang, MD, PhD, Ying Wang, MD, Mei Lu, PhD, Margot LaPointe, PhD, and Michael Chopp, PhD

612 **Human Herpesvirus 6 Limbic Encephalitis After Stem Cell Transplantation**
Mark S. Wainwright, MD, PhD, Paul L. Martin, MD, PhD, Richard P. Morse, MD, Mary Lacaze, MD, James M. Provenzale, MD, R. Edward Coleman, MD, Marcello A. Morgan, Christine Hulette, MD, Joanne Kurtzberg, MD, Cheryl Bushnell, MD, Leon Epstein, MD, and Darrell V. Lewis, MD

620 **Plasticity of Language Networks in Patients with Brain Tumors: A Positron Emission Tomography Activation Study**
Alexander Thiel, MD, Karl Herholz, MD, Adem Koyuncu, Mehran Ghaemi, MD, Lutz W. Kracht, MD, Birgit Habedank, and Wolf-Dieter Heiss, MD

630 **Induction of Proinflammatory Molecules in Mice with Amyotrophic Lateral Sclerosis: No Requirement for Proapoptotic Interleukin-1β in Neurodegeneration**
Minh Dang Nguyen, BSc, Jean-Pierre Julien, PhD, and Serge Rivest, PhD

640 **Depletion of Corticotrophin-Releasing Factor Neurons in the Pontine Micturition Area in Multiple System Atrophy**
Eduardo E. Benarroch, MD, DSci, and Ann M. Schmeichel, AS

646 **Immunopathology of Secondary-Progressive Multiple Sclerosis**
John W. Prineas, MB, BS, Eunice E. Kwon, MSc, Eun-Sook Cho, MD, Leroy R. Sharer, MD, Michael H. Barnett, MB, BS, Emilia L. Oleszak, PhD, Brad Hoffman, BS, and Bryan P. Morgan, PhD

Brief Communications

658 **Case-Control Study of the Extended Tau Gene Haplotype in Parkinson's Disease**
Demetrius M. Maraganore, MD, Dena G. Hernandez, BS, Andrew B. Singleton, PhD, Matthew J. Farrer, PhD, Shannon K. McDonnell, MS, Michael L. Hutton, PhD, John A. Hardy, PhD, and Walter A. Rocca, MD, MPH

661 **Endocytic Disturbances Distinguish Among Subtypes of Alzheimer's Disease and Related Disorders**
Anne Cataldo, PhD, G. William Rebeck, PhD, Bernadino Ghetti, MD, Christine Hulette, MD, Carol Lippa, MD, Christine Van Broeckhoven, PhD, DrSc, Cornelia van Duijn, PhD, Patrick Cras, MD, PhD, Nenad Bogdanovic, MD, Thomas Bird, MD, Corrinne Peterhoff, BS, and Ralph Nixon, MD, PhD

665 **Adult-Onset Leukoencephalopathy with Vanishing White Matter Presenting with Dementia**
Konstantin Prass, MD, Wolfgang Brück, MD, Nicolas W. J. Schröder, Andreas Bender, MD, Maria Prass, MD, Tilo Wolf, MD, Marjo S. Van der Knaap, MD, PhD, and Rolf Zschenderlein, MD

668 **Relapsing–Remitting Autoimmune Agrypnia**
Anna Paola Batocchi, MD, Giacomo Della Marca, MD, Massimiliano Mirabella, MD, Marcella Caggiula, MD, Giovanni Frisullo, MD, Gioacchino Francesco Mennuni, MD, and Pietro Attilio Tonali, MD

672 **Functional Activation of Microgyric Visual Cortex in a Human**
Giorgio M. Innocenti, MD, Philippe Maeder, MD, Maria G. Knyazeva, PhD, Eleonora Fornari, and Thierry Deonna, MD

677 **Convergence Retraction Nystagmus: A Disorder of Vergence?**
Holger Rambold, MD, Detlef Kömpf, MD, and Christoph Helmchen, MD

681 **Dysautonomia in Human T-Cell Lymphotrophic Virus Type I-Associated Myelopathy/Tropical Spastic Paraparesis**
Alexandre H. Alamy, MD, MSc, Fernando B. Menezes, MD, MSc, Ana C. B. Leite, MD, PhD, Osvaldo M. Nascimento, MD, PhD, and Abelardo Q-C. Araújo, MD, PhD

Letters

686 **Helicobacter Pylori-Induced Reduction of Acute Levodopa Absorption in Parkinson's Disease Patients**
Mariangela Pierantozzi, PhD, Antonio Pietroiusti, MD, Alberto Galante, MD, Giuseppe Sancesario, MD, Gianluigi Lunardi, PhD, Ernesto Fedele, PhD, Patrizia Giacomini, MD, and Paolo Stanzione, MD

687 **Pharmacodynamic Modeling of Oral Levodopa in Parkinson's Disease**
Manuela Contin, PharmD, Roberto Riva, MD, Fiorenzo Albani, PharmD, Paolo Martinelli, MD, and Agostino Baruzzi, MD

688 **Reply**
Raúl de la Fuente-Fernández, MD, and A. Jon Stoessl, MD, FRCPC

688 **Are the "Newly Discovered" Paraneoplastic Anti-Collapsin Response-Mediator Protein 5 Antibodies Simply Anti-CV2 Antibodies?**
Jérôme Honnorat, MD, PhD, Jean-Christophe Antoine, MD, and Marie-Françoise Belin, PhD

690 **Reply**
Vanda A. Lennon, MD, PhD, Zhiya Yu, MD, PhD, Thomas J. Kryzer, AS, and Guy E. Griesmann, MS

692 **Classic Rett Syndrome in a Boy as a Result of Somatic Mosaicism for a *MECP2* Mutation**
Judith Armstrong, MsC, Pilar Póo, MD, PhD, Mercè Pineda, MD, PhD, Elena Aibar, MsC, Esther Geán, MsC, MD, Vicenç Català, MsC, PhD, and Eugènia Monrós, MsC, PhD

Regular Departments

693 **Books**

NO 6 DECEMBER 2001

Editorials

695 **Microarrays, Markers of Disease, and the Myth of "Nonhypothesis-Driven" Research**
David A. Greenberg, MD, PhD

696 **Epilepsy After Head Injury: The Impact of Impact**
Daniel Lowenstein, MD

697 **Imaging Stroke Recovery: Lessons from Prozac**
John A. Detre, MD

Original Articles

699 **Blood Genomic Responses Differ After Stroke, Seizures, Hypoglycemia, and Hypoxia: Blood Genomic Fingerprints of Disease**
Yang Tang, MD, Aigang Lu, MD, Bruce J. Aronow, PhD, and Frank R. Sharp, MD

708 **Long-Term Hyperexcitability in the Hippocampus After Experimental Head Trauma**
Vijayalakshmi Santhakumar, BS, Anna D. H. Ratzliff, BS, Jade Jeng, BS, Zsolt Toth, MD, PhD, and Ivan Soltesz, PhD

718 **Fluoxetine Modulates Motor Performance and Cerebral Activation of Patients Recovering from Stroke**
Jérémie Pariente, MD, Isabelle Loubinoux, PhD, Christophe Carel, MD, Jean-François Albucher, MD, Anne Leger, MD, Claude Manelfe, MD, PhD, Olivier Rascol, MD, PhD, and François Chollet, MD

730 **Disease Mechanisms Revealed by Transcription Profiling in SOD1-G93A Transgenic Mouse Spinal Cord**
Mary K. Olsen, BA, Steven L. Roberds, PhD, Brenda R. Ellerbrock, BA, Timothy J. Fleck, BA, Denise K. McKinley, BA, and Mark E. Gurney, PhD

741 **Dementia and Neurodevelopmental Predisposition: Cognitive Dysfunction in Presymptomatic Subjects Precedes Dementia by Decades in Frontotemporal Dementia**
Daniel H. Geschwind, MD, PhD, Janik Robidoux, PhD, Maricela Alarcón, PhD, Bruce L. Miller, MD, Kirk C. Wilhelmsen, MD, PhD, Jeffrey L. Cummings, MD, and Ziad S. Nasreddine, MD

747 **Compensating for Central Nervous System Dysmyelination: Females with a Proteolipid Protein Gene Duplication and Sustained Clinical Improvement**
Ken Inoue, MD, PhD, Hajime Tanaka, MD, PhD, Fernando Scaglia, MD, Akiko Araki, MD, Lisa G. Shaffer, PhD, and James R. Lupski, MD, PhD

755 **McLeod Neuroacanthocytosis: Genotype and Phenotype**
Adrian Danek, MD, PhD, Justin P. Rubio, PhD, Luca Rampoldi, PhD, Mengfatt Ho, D Phil (Oxon), Carol Dobson-Stone, MBiochem, François Tison, MD, PhD, William A. Symmans, MD, Matthias Oechsner, MD, Wolfgang Kalckreuth, MD, Julie M. Watt, BSc, Alastair J. Corbett, MD, FRACP, Hisham H. M. Hamdalla, MD, MRCP (UK), Andrew G. Marshall, MD, MRCP (UK), Ian Sutton, MD, MRCP (UK), Maria Teresa Dotti, MD, Alessandro Malandrini, MD, Ruth H. Walker, MD, PhD, Geoff Daniels, PhD, and Anthony P. Monaco, MD, PhD

765 **Dementia in Hereditary Cerebral Hemorrhage with Amyloidosis-Dutch Type Is Associated with Cerebral Amyloid Angiopathy but Is Independent of Plaques and Neurofibrillary Tangles**
Remco Natté, MD, Marion L. C. Maat-Schieman, MD, PhD, Joost Haan, MD, PhD, Marjolijn Bornebroek, MD, PhD, Raymund A.C. Roos, MD, PhD, and Sjoerd G. van Duinen, MD, PhD

773 **The Wld^S Protein Protects Against Axonal Degeneration: A Model of Gene Therapy for Peripheral Neuropathy**
Min Sheng Wang, PhD, Guofu Fang, MS, Deborah G. Culver, MS, Albert A. Davis, Mark M. Rich, PhD, MD, and Jonathan D. Glass, MD

780 **Cigarette Smoking and the Incidence of Parkinson's Disease in Two Prospective Studies**
Miguel A. Hernán, MD, Shumin M. Zhang, MD, Ana M. Rueda-deCastro, BS, Graham A. Colditz, MD, Frank E. Speizer, MD, and Alberto Ascherio, MD

787 **Effect of Therapeutic Ionizing Radiation on the Human Brain**
R. Grant Steen, PhD, David Spence, BS, Shengjie Wu, MS, Xiaoping Xiong, PhD, Larry E. Kun, MD, and Thomas E. Merchant, DO, PhD

Brief Communications

796 **Molecular and Clinical Correlation in Five Indian Families with Spinocerebellar Ataxia 12**
Achal K. Srivastava, DM, Shweta Choudhry, MSc, Musuwadi S. Gopinath, MD, Sanghamitra Roy, MSc, Manjari Tripathi, DM, Samir K. Brahmachari, PhD, and Satish Jain, DM

800 **Reorganization of Somatic Sensory Function in the Human Thalamus After Stroke**
Shinji Ohara, MD, PhD, and Frederick A. Lenz, MD, PhD

804 **Evidence for Postjunctional Serotonin (5-HT$_1$) Receptors in the Trigeminocervical Complex**
Peter J. Goadsby, MD, DSc, Simon Akerman, MSc, and R. James Storer, PhD

807 **Increased HTLV-I Proviral Load and Preferential Expansion of HTLV-I Tax-Specific CD8 T Cells in Cerebrospinal Fluid from Patients with HAM/TSP**
Masahiro Nagai, MD, PhD, Yoshihisa Yamano, MD, PhD, Meghan B. Brennan, BA, Carlos A. Mora, MD, and Steven Jacobson, PhD

812 **Spinocerebellar Ataxia Type 2 Presenting as Familial Levodopa-Responsive Parkinsonism**
Din-E Shan, MD, PhD, Bing-Wen Soong, MD, PhD, Chen-Ming Sun, MD, Shwn-Jen Lee, PhD, PT, Kwong-Kum Liao, MD, and Ren-Shyan Liu, MD

816 **Complete Allele Information in the Diagnosis of Facioscapulohumeral Muscular Dystrophy by Triple DNA Analysis**
Richard J. L. F. Lemmers, BSc, Peggy de Kievit, Michel van Geel, PhD, Michiel J.R. van der Wielen, Egbert Bakker, PhD, George W. Padberg, MD, PhD, Rune R. Frants, PhD, and Silvère M. van der Maarel, PhD

820 **So Near Yet So Far: Neglect in Far or Near Space Depends on Tool Use**
Alan J. Pegna, PhD, Leila Petit, MA, Anne-Sarah Caldara-Schnetzer, MA, Asaid Khateb, PhD, Jean-Marie Annoni, MD, Roman Sztajzel, MD, and Theodor Landis, MD

Letters

823 **N-Acetylaspartate—A Marker of Neuronal Integrity**
Edith V. Sullivan, PhD, Elfar Adalsteinsson, PhD, Daniel M. Spielman, PhD, Ralph E. Hurd, PhD, and Adolf Pfefferbaum, MD

823 **N-Acetylaspartate: Usefulness as an Indicator of Viable Neuronal Tissue**
Douglas L. Arnold, Nicola de Stefano, Paul M. Matthews, and Bruce D. Trapp

824 **Reply**
Ernst Martin, MD, Thorsten Thiel, PhD, Andrea Capone, MD, Juergen Henning, PhD, and Jacques Schneider, MD

825 **Bilateral High-Frequency Synchronous Discharges Associated with Posterior Fossa Disorders**
Mario-Ubaldo Manto, MD, and John Moore, BE

827 **Effectiveness of Intravenous Immunoglobin Therapy in Cerebellar Ataxia Associated with Gluten Sensitivity**
Katrin Bürk, MD, Arthur Melms, MD, Jörg B. Schulz, MD, and Johannes Dichgans, MD

828 **Cerebrospinal Fluid Interleukin-6 Levels: A Possible Marker of Disease Activity**
Kurt A. Jellinger, MD, PhD, Norbert Rösler, MD, and Ildiko Wichart, PhD

829 **Delayed Synaptic Dysfunction of Association Cortices in Carbon Monoxide Intoxication**
Masafumi Ihara, MD, Hidenao Fukuyama, MD, PhD, Tetsuya Lee, MD, Sumiko Takao, MD, Nobuo Kohara, MD, PhD, and Hiroshi Shibasaki, MD, PhD

Obituary

831 **Maurice Victor: 1920–2001**
Richard T. Johnson, MD

Regular Departments

830 **Corrections**
832 **Books**
834 **American Neurological Association, Works in Progress Abstracts**

Indexes to Volume 50

837 **Author Index**
847 **Subject Index**